CASE REPORT

Oesophageal lesions as first manifestation of necrotising sarcoid granulomatosis

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SUMMARY

Necrotising sarcoid granulomatosis is a rare disease associating sarcoid-like granulomas, vasculitis and necrosis in the lungs and seldom in extrapulmonary locations. We report the case of a 23-year-old woman with an initial symptom consisting of epigastric pain. Oesophagoscopy demonstrated oesophagitis. Evolution was marked by fever, dyspnoea and pulmonary lesions. The first presumptive diagnosis was Wegener disease but biopsies (of the lungs and the oesophagus) demonstrated the diagnosis of necrotising sarcoid granulomatosis. Evolution was favourable under corticoid therapy. Necrotising sarcoid granulomatosis may present as an extrapulmonary disease and is an important differential diagnosis of other disease such as Wegener disease.

BACKGROUND

Necrotising sarcoid granulomatosis (NSG) affects young or middle-aged participants and has a slight female preponderance. The symptoms are non-specific and consist of cough, fever, fatigue, chest pain and dyspnoea. Since its description by Liebow¹, a 100 cases of NSG have been reported in the literature. This is the first reported case of upper digestive tract lesions as the first manifestation of the disease.

CASE PRESENTATION

A 23-year-old woman sought advice from her family doctor for epigastric pain and vomiting for a week. Her medical history was unremarkable except for childhood allergic asthma. She had no known immunodeficiency. Her clinical examination was normal. An oesophagogastroscope showed grade C oesophagitis and gastritis with positive *Helicobacter pylori* culture. Evolution was unfavourable despite treatment with pantoprazole and clarithromycin. A week later, the patient was admitted to the emergency department with fever (38°C), cough and chest pain radiating to the left shoulder. Laboratory tests showed an inflammatory syndrome with a C reactive protein level at 6.3 mg% (normal range <0.5), leucocytosis at 15.800/mm³ (4–12 000) and mild disturbances of liver function. Chest radiography revealed basal opacities of both lungs (figure 1A). Atypical pneumonia was suspected and the patient was sent home with moxifloxacin for 10 days. She was readmitted on day 10 with persistent fever and onset of dyspnoea. On admission, the physical examination, especially pulmonary auscultation, was normal except for fever. Laboratory tests confirmed earlier findings

(inflammatory syndrome and liver enzymes abnormalities). Antinuclear factor, antineutrophil cytoplasmic antibodies, rheumatoid factor and Waaler-Rose were all negative. Angiotensin convertase levels were normal. All bacteriological and serological tests were negative. Urinalysis was normal. Functional pulmonary testing revealed a severe restrictive lung disease (forced expiratory volume 37% of predicted value, forced vital capacity ratio of 91% and transfer coefficient for carbon monoxide of 31%) with diffusion alterations. Bronchoscopy showed no endobronchial lesions, and bacteriological and cytological samples were negative. Cerebral MRI and funduscopy showed no signs of vasculitis.

Thoracic tomodensitometry (figure 1B) showed multiple pulmonary nodules and confluent areas of condensation, especially in the lower lobes. In the periphery of these lesions, ground-glass opacities and nodular thickening of the caesuras were seen. No mediastinal or hilar lymphadenopathy was present. Surgical lung biopsy was performed and provided a definitive diagnosis of NSG with the association of granulomas, vasculitis and areas of necrosis (figure 2). Oesophageal biopsy showed the same histological findings.

INVESTIGATIONS

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DIFFERENTIAL DIAGNOSIS

Lung biopsy excluded the presumptive diagnosis of Wegener's disease.

TREATMENT

The patient's clinical course was rapidly favourable under corticosteroids.

OUTCOME AND FOLLOW-UP

Follow-up was unremarkable, with corticosteroids weaned after 12 months. Functional pulmonary testing remained close to normal (diffusion slightly decreased) at 2-year follow-up.

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Figure 1 (A) Chest X-ray: bibasilar ill-defined nodular opacities. (B) Thoracic CT: multiple and bibasilar coalescent pulmonary nodules and consolidations (asterisk), surround by patchy ground-glass opacities (black arrow). Nodular thickening of major fissure (white arrow).

Figure 2 Anatomopathological findings of the lung. (A) Figure showing granulomas in the lungs (H&E stain, $\times 40$). (B) Figure showing a vessel (Orcein staining, $\times 20$) affected by a chronic vasculitis (brown) surrounded by granulomas. The Periodic acid–Schiff and Ziehl stainings did not show any microorganisms.

DISCUSSION

Of primary interest in our observation were the exceptional upper gastrointestinal tract lesions (to the best of our knowledge this is the first case described).

The second significant feature was the difficulty in establishing a differential diagnosis with other pathologies such as Wegener's granulomatosis, and the therapeutic and prognostic implications that this major differential diagnosis implies. Indeed, in mild Wegener disease, methotrexate is often associated with corticosteroids, and in severe disease,

cyclophosphamide is associated with rituximab, which can lead to possible severe side effects.

The radiological findings in NSG are solitary or bilateral, multiple nodules, with possible cavitation, alveolar infiltrates with the possible presence of hilar or mediastinal adenopathies on chest CT.²

It is critical to histopathologically exclude all other possible infectious and non-infectious diseases to arrive at the correct diagnosis (box 1). Extra-respiratory manifestations of NSG include ophthalmological, neurological and, extremely rarely, digestive^{2–4} findings. The relationship between NSG and sarcoidosis is controversial:^{2 3 5} some have suggested NSG as a

Box 1 Differential diagnosis of pulmonary granulomatous diseases

Infections: mycobacteria (tuberculous and non-tuberculous), fungi (*Aspergillus*, *Cryptococcus*, *Histoplasma*, *Pneumocystis jiroveci*, etc) parasites (*Dirofilaria*)

- ▶ Hypersensitivity pneumonia
- ▶ Wegener granulomatosis
- ▶ Churg-Strauss syndrome
- ▶ Lymphoid interstitial pneumonia
- ▶ Pneumoconiosis (Beryllium)
- ▶ Talc granulomatosis
- ▶ Langerhans cell histiocytosis
- ▶ Sarcoidosis
- ▶ Necrotising sarcoid granulomatosis

Learning points

- ▶ Necrotising sarcoid granulomatosis is an infrequent disease, especially in its gastrointestinal form.
- ▶ The diagnosis may be challenging and needs histopathological evidence to exclude other granulomatous diseases, as it is indispensable to ensure optimal treatment. Clinicians must be aware of this rare but clinically important disease.
- ▶ In our case, the initial symptoms and lesion localisation were oesophageal, which has never been described before.
- ▶ The evolution is usually benign but must be treated by corticosteroids in the event of systemic disease.

variant of sarcoidosis, but the specific histopathological data are probably sufficient to suggest that NSG is a separate entity. One must bear in mind that in sarcoidosis, oesophageal involvement is described but extremely rare.⁶

The prognosis of NSG is usually favourable; spontaneous resolution is possible and treatment with corticosteroids (or in a minority of cases, other immunosuppressive drugs) should be restricted to those patients with major symptoms.²

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Liebow AA. The J. Burns Amberson lecture—pulmonary angiitis and granulomatosis. *Am J Respir Dis* 1973;108:1–18.
- 2 Quaden C, Tillie-Leblond I, Delobbe A, *et al*. Necrotising sarcoid granulomatosis: clinical, functional, endoscopic and radiographical evaluations. *Eur Respir J* 2005;26:778–85.
- 3 Le Gall F, Loeuillet L, Delaval P, *et al*. Necrotizing sarcoid granulomatosis with and without extrapulmonary involvement. *Pathol Res Pract* 1996;192:306–14.
- 4 Dykhuizen RS, Smith CC, Kennedy MM, *et al*. Necrotizing sarcoid granulomatosis with extrapulmonary involvement. *Eur Respir J* 1997;10:245–7.
- 5 Popper HH, Klemen H, Colby TV, *et al*. Necrotizing sarcoid granulomatosis—is it different from nodular sarcoidosis? *Pneumologie* 2003;57:268–71.
- 6 Abraham A, Hajar R, Virdi R, *et al*. Esophageal sarcoidosis: a review of cases and an update. *ISRN Gastroenterol* 2013;2013:836203.

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